Concise summary of the 2012 World Heart Federation criteria for the echocardiographic diagnosis of rheumatic heart disease

Congenital, acquired and degenerative heart disease should always be excluded as the etiology of MV and AV abnormalities. Echocardiographic features should be interpreted in conjunction with other indicators of risk for RHD (e.g. demographics and regional differences) and clinical findings.

Echocardiographic criteria for individuals aged ≤20 years

Definite RHD (either A, B, C, or D):

A) Pathological MR and at least two morphological features of RHD of the MV
B) MS mean gradient ≥4 mmHg*
C) Pathological AR and at least two morphological features of RHD of the AV‡
D) Borderline disease of both the AV and MV§

Borderline RHD (either A, B, or C):

A) At least two morphological features of RHD of the MV without pathological MR or MS
B) Pathological MR
C) Pathological AR

Normal echocardiographic findings (all of A, B, C, and D);

A) MR that does not meet all four Doppler echocardiographic criteria (physiological MR)
B) AR that does not meet all four Doppler echocardiographic criteria (physiological AR)
C) An isolated morphological feature of RHD of the MV (e.g. valvular thickening) without any associated pathological stenosis or regurgitation
D) Morphological feature of RHD of the AV (e.g. valvular thickening) without any associated pathological stenosis or regurgitation

Echocardiographic criteria for individuals aged >20 years

Definite RHD (either A, B, C, or D):

A) Pathological MR and at least two morphological features of RHD of the MV
B) MS mean gradient ≥4 mmHg*
C) Pathological AR and at least two morphological features of RHD of the AV, only in individuals aged <35 years‡
D) Pathological AR and at least two morphological features of RHD of the MV

Echocardiographic criteria for pathological regurgitation

Pathological MR

(All 4 Doppler echocardiographic criteria must be met)

1. Seen in two views
2. In at least one view, jet length ≥2 cm‖
3. Velocity ≥3 m/s for one complete envelope
4. Pan-systolic jet in at least one envelope

Pathological AR

(All 4 Doppler echocardiographic criteria must be met)

1. Seen in two views
2. In at least one view, jet length ≥1 cm‖
3. Velocity ≥3 m/s in early diastole
4. Pan-diastolic jet in at least one envelope

Morphological features of RHD

Features in the MV

- AMVL thickening¶ ≥3 mm (age-specific)#
- Chordal thickening
- Restricted leaflet motion**
- Excessive leaflet tip motion during systole‡‡

Features in the AV

- Irregular or focal thickening§§
- Coaptation defect
- Restricted leaflet motion
- Prolapse

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Important considerations

*Congenital MV anomalies must be excluded. Furthermore, inflow obstruction due to nonrheumatic mitral annular calcification must be excluded in adults.

‡Bicuspid AV, dilated aortic root, and hypertension must be excluded.

§Combined AR and MR in high prevalence regions and in the absence of congenital heart disease is regarded as rheumatic.

‖A regurgitant jet length should be measured from the vena contracta to the last pixel of regurgitant colour (blue or red)

¶AMVL thickness should be measured during diastole at full excursion. Measurement should be taken at the thickest portion of the leaflet, including focal thickening, beading, and nodularity. Measurement should be performed on a frame with maximal separation of chordae from the leaflet tissue. Valve thickness can only be assessed if the images were acquired at optimal gain settings without harmonics and with a frequency ≥2.0 MHz.

#Abnormal thickening of the AMVL is age-specific and defined as follows: ≥3 mm for individuals aged ≤20 years; ≥4 mm for individuals aged 21–40 years; ≥5 mm for individuals aged >40 years. Valve thickness measurements obtained using harmonic imaging should be cautiously interpreted and a thickness up to 4 mm should be considered normal in those aged ≤20 years.

**Restricted leaflet motion of either the anterior or the posterior MV leaflet is usually the result of chordal shortening or fusion, commissural fusion, or leaflet thickening.

‡‡Excessive leaflet tip motion is the result of elongation of the primary chords, and is defined as displacement of the tip or edge of an involved leaflet towards the left atrium resulting in abnormal coaptation and regurgitation. Excessive leaflet tip motion does not need to meet the standard echocardiographic definition of MV prolapse disease, as that refers to a different disease process. This feature applies to only those aged <35 years. In the presence of a flail MV leaflet in the young (≤20 years), this single morphological feature is sufficient to meet the morphological criteria for RHD (that is, where the criteria state "at least two morphological features of RHD of the MV" a flail leaflet in a person aged ≤20 years is sufficient).

§§In the parasternal short axis view, the right and noncoronary aortic cusp closure line often appears echogenic (thickened) in healthy individuals and this should be considered as normal.

Echocardiography machine settings

- Nyquist limits for colour-Doppler echocardiography should be set on maximum to avoid overestimation of jet length.
- Images for assessment of valvular and chordal thickness should be acquired with harmonics turned off and probes with variable frequency set on ≥2.0 MHz. Low frequency settings and harmonics exaggerate valve and chordal thickness.
- Gain settings should be adjusted to achieve optimal resolution. Images acquired with an excessive gain setting will not be suitable for objective valve thickness measurements.
- All other settings (including depth, sector size, and focus) should also be optimized to achieve maximal frame rate (ideally 30–60 frames per second) and resolution.

Citation for the full version of these guidelines: Reményi, B. *et al*. World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease—an evidence-based guideline. *Nat. Rev. Cardiol.* **9**, 297–309 (2012).

Abbreviations: AR, aortic regurgitation; AMVL, anterior mitral valve leaflet; AV, aortic valve; MR, mitral regurgitation; MS, mitral stenosis; MV, mitral valve; RHD, rheumatic heart disease.

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